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Epispadias with complete prepuce: A rare anomaly

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Epispadias with complete prepuce is a very rare anomaly. It is often associated with late presentation because it is a very rare condition and the penis appears grossly normal, thus, the diagnosis is easily missed during the neonatal period. We report a case of a boy presenting at seven years of age with epispadias and complete prepuce. The diagnosis should be suspected in any child presenting with incontinence, short penis, ballooned prepuce on voiding and absence of penile raphe or frenulum. The established procedures for repair of classic epispadias are equally effective and bladder neck reconstruction may be required to achieve continence.

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Introduction

Epispadias with complete prepuce is a very rare anomaly. It is often associated with late presentation because the diagnosis can easily be missed during the neonatal period. Because of the rarity of the condition, the incidence is not known but a number of cases have been reported in literature. We report a case of a boy presenting at

seven years of age with epispadias and complete prepuce. We also did a review of literature on the clinical presentation and surgical management of the condition.

Case summary

A 7 year old boy presented to our Urology clinic with continuous dribbling of urine since birth, however he also voids with a stream associated with ballooning of the penis from time to time, the penis was said to be straight on erection and not painful. There was no history of other anomalies and previous operations. He is a product of term uneventful gestation and vaginal delivery. On examination he was a well preserved male child, with normal gait. Examination of the external genitalia revealed a short conically shaped penis with complete prepuce, the preputial opening was stenosed and dorsally directed (Fig. 1), the preputial skin ballooned out on attempt at micturition (Fig. 2). There was a palpable dorsal groove on palpation. He had a well developed scrotum containing normal size testes. Gentle retraction of the prepuce, revealed a broad spade like glans and penile epispadias, there was mild dorsal chordee with obvious

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Figure 1 A conically shaped short penis with complete prepuce.

urinary incontinence (Fig. 3). A diagnosis of penile epispadias with complete prepuce was made.

Abdominopelvic ultrasound and other baseline investigations were normal. Micturating cystourethrogram (MCUG) showed intact pelvic bones with no pubic symphyseal diasthesis on the plain film. The contrast study revealed a good bladder capacity without vesicoureteric reflux, the posterior urethra was normal and the preputial skin was ballooned with contrast (Fig. 4).

At repair a circumcising incision was made and the penis degloved to reveal the entire extent of the defect. An inverted U shaped incision was made around the urethral plate without complete mobilization of the plate. The urethral plate was closed over a 10 French tube (Byar's urethroplasty) and the corporal bodies approximated over the reconstructed urethra. Chordee was surprisingly mild and did not require correction, thus induction of artificial erection was not required. Skin cover was easily achieved with the abundant circumferential penile skin, suprapubic urinary diversion was done (Fig. 5a and b). Postoperatively, he voided with good stream after removal of the urethral stent on the 4th day and suprapubic drain was removed



Figure 2 A dorsally directed pretucial opening and ballooning of the prepuce on micturition.

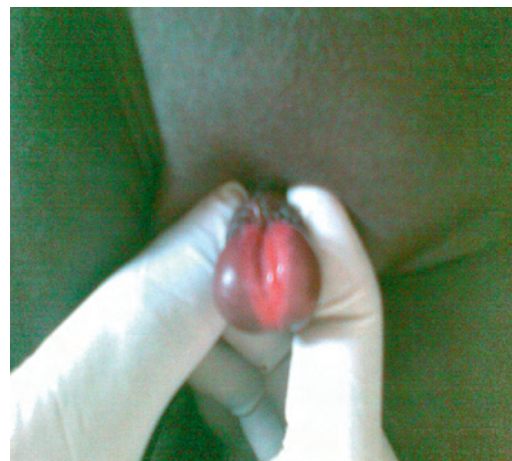


Figure 3 Penile epispadias on preputial retraction.

on the 7th day. Patient was followed up in the outpatient clinic at intervals and at three months post repair, he was still dribbling urine between good volitional voids. The persistent incontinence was to be addressed by a later bladder neck reconstruction.

Discussion

Isolated epispadias is a rare anomaly with a reported incidence of 1 in 117 000 males [1], epispadias is classically associated with bladder extrophy in over 90% of cases [2], isolated epispadias without extrophy is thus less common. The prepuce is usually deficient on the dorsal aspect of the penis in the typical epispadias, the presence of a complete prepuce associated with epispadias as in our case is extremely rare but has been described in some patients [1–5].

It is often associated with late presentation because the diagnosis can be missed during the neonatal period [4] due to the apparent normal appearance of the penis. This is because the complete prepuce conceals the penile anomaly and preclude early detection, therefore these patients may only present when it is discovered at circumcision (where routine religious/cultural circumcision is practiced), when the child is noticed to have an abnormally short penis for his age or during evaluation for incontinent of urine (when the parents



Figure 4 MCUG with ballooned prepuce.

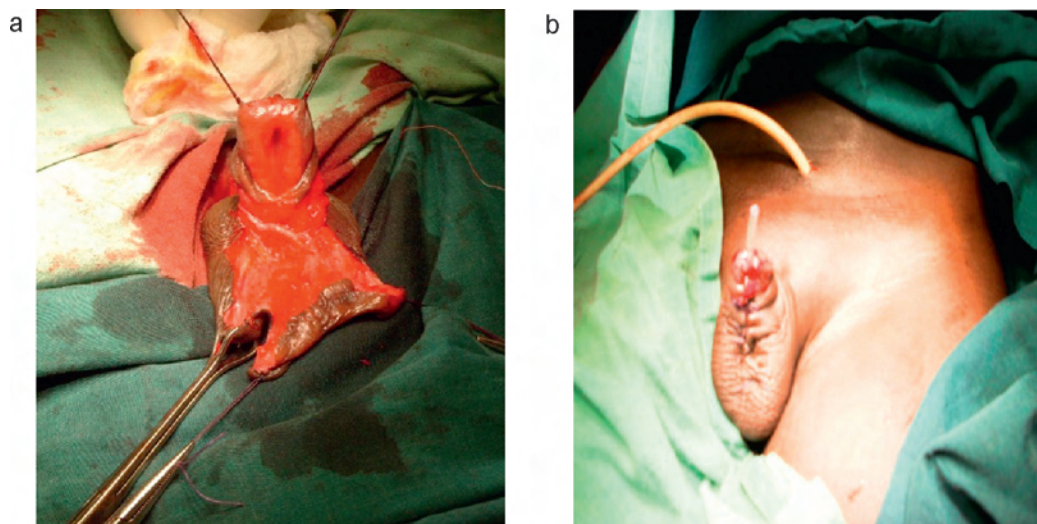


Figure 5 (a) Degloved penis showing a penile epispadias and (b) appearance of penis after completion of repair.

complain of persistent nocturnal enuresis or continuous dribbling). The clinical signs that may raise the suspicion of this condition have been described in literature [1,5]; a short penis with a broad spade like glans, absence of frenulum, and penile raphe on the glans and a dorsally directed preputial opening. A dorsal midline depression with separation of the corporal bodies could be felt on palpation. Our patient presented at the age of seven years and the chief complaint of the parents was incontinence. The presence of a complete prepuce, a moderate sized penis and the absence of significant dorsal chordee contributed to the late presentation in this case.

There has been no satisfactory explanation for presence of a complete prepuce in an epispadic penis, this is made worse by the extreme rarity of the condition. Most of the few reported cases were found among patients with urethral defect restricted to the glans penis, this prompted its comparison with the 'mega urethra intact prepuce' found in some hypospadias. Raghavaiah suggested that in the case of glandular epispadias, the preputial development can progress to completion since the prepuce usually originates from a fold of ectoderm at the level of the coronal sulcus [6]. However, this could not explain the cases of epispadias with urethral defect proximal to the coronal sulcus with complete prepuce (as in the index patient we are reporting). Merlob et al. felt the occurrence of this rare condition is a complete deviation from normal development of the external genitalia rather than a mere developmental arrest, they also stated that the presence of a dorsally directed preputial opening, the absence of frenulum on the glans and incomplete penile raphe are further evidence of an abnormal development [2].

Surgical repair can significantly correct the anomaly with acceptable functional and cosmetic outcome. Any of the procedures described for the repair of typical epispadias is applicable and for the glandular type a simple glans approximation with or without circumcision may suffice. The advantage we noticed at surgery in this patient with a complete prepuce is the abundance of skin for easy closure giving a better cosmetic appearance. Though the epispadias in this case was penile and the patient was able to achieve intermittent volitional voids, the persistence of dribbling postoperatively suggests an inherent subtle degree of incontinence. The adequate to normal capacity appearance of the urinary bladder on the MCUG

(Fig. 4) suggests that the patient is able to retain urine enough for voluntary micturition, thus, ruling out complete incontinence. Distal epispadias (penile and glandular) are commonly not associated with incontinence; however, some penile epispadias have been observed to exhibit variable degrees of incontinence. In a review by Culp in 1973, of the 6 patients with penile epispadias 3 were incontinent and 3 had an element of continence [7]. Thus a simple repair without continence surgery will probably not suffice in some patients. To address the incontinence in our patient, the parents were counseled about the possibility of bladder neck reconstruction if the child remained incontinent.

Conclusion

The diagnosis of this condition is often missed in the neonatal period, thus patients tend to present late. A high index of suspicion is required to make the diagnosis, it should be suspected in any child presenting with incontinence, short penis, ballooned prepuce on voiding and absence of penile raphe or frenulum. The established procedures for repair of classic epispadias are equally effective and bladder neck reconstruction may be required to achieve continence.

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